

REMARKS ON PRIMARY NEUROTIC ATROPHY (CHARCOT-MARIE-HOFFMAN TYPE), WITH REPORT OF A CASE
IN WHICH THERE WAS EXCESSIVE INDULGENCE IN TEA AND COFFEE.¹

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The clinical varieties of progressive muscular atrophy are not as yet sharply defined. Within the last few years a number of facts have accumulated, showing the difficulty of differentiating distinctly the types to which certain forms of muscular atrophy belong. Symptoms which were considered characteristic of one form are now found to be present also in another, as for example, the reactions of degeneration, fibrillary twitching, or the mode of development of the atrophy.

The progressive muscular atrophy described by Charcot and Marie, and later by Hoffman, presents a variety which is difficult to classify, as the pathological findings are not identical in every case. The clinical picture of this form presents partly symptoms of Aran-Duchenne's disease, and partly symptoms of myopathy. However, owing to the fact that the initial symptoms are in the majority of cases identical, namely, atrophy of the peroneal group of muscles, the disease can be considered as a separate form. Moreover, if the spinal changes have not always been found to be absolutely identical, changes in the peripheral nerves have been observed in the majority of cases. The name neurotic or neuritic atrophy is justified, but it is doubtful whether the nature of the affection is always primarily in the peripheral nerves.

In cases which present spinal changes in addition to those of the peripheral nerves, it is extremely difficult to determine which of the two was the initial lesion. For this reason, the term spinal-neuritic atrophy would be more appropriate. In fact, the clinical picture of the majority of cases shows an almost simultaneous onset of the atrophy in the lower extremities and in the small muscles of the hand, thus imitating the picture of the Aran-Duchenne's type for the upper extremities. Charcot and Marie did not believe in the neuritic origin of their disease, but the

¹This paper was read and the patient was exhibited before the Philadelphia Neurological Society, February 24, 1903.

cases reported later show by microscopical examination that the disease is both myelopathic and neuritic. The few cases recorded with total absence of spinal changes, or with very slight changes in the cord (Dubreuil²) do not, in my opinion, exclude the possibility of a spinal involvement, as it is probable that the patients did not live long enough for the spinal changes to develop.

The case I am about to report is an example of this spinal-neuritic form of progressive muscular atrophy. The particular interest lies in the associated symptoms and in the history. It also shows the difficulty in classification.

Mr. S. T., aged thirty-seven, gives the following history: For twenty years he had been in charge of a grocery store, where he worked daily from 5 A. M. to 12 P. M. He was constantly exposed to cold. In the winter of 1897, for the first time he felt pain in the toes of both feet. The pain was accompanied by a burning sensation. The suffering at times was so intense that he had to give up work. Application of heat would make the condition worse. For four and one-half years the only symptom was pain.

Three months ago he noticed that in walking he had to raise his feet very high, and that he could not flex the foot upon the leg, especially on the right side.

Status praesens:—Inspection shows the legs emaciated, especially in the lower halves. The muscles of the thighs are also somewhat wasted, and the adductors are flabby. The calf muscles of both legs, and especially those of the right, are soft and flabby and give the impression of the pseudo-hypertrophic type of myopathy. The gluteal muscles are flabby. The feet are cyanosed and present the equino-varus type of club-foot. The patient stands on his metatarso-phalangeal articulations, the second and third phalanges are markedly bent downwards. The foot-drop is more marked on the right side than on the left. The nails have suffered considerably in their nutrition; some of them are absent and some are retracted.

The motor apparatus shows the following disturbances: Gait is ataxic and presents the typical high steppage, especially in the right limb. Station with closed eyes is only somewhat impaired. The knee-jerks are totally absent, even with reinforcement. The tendon of Achilles reflex is also absent on each side. No ankle-clonus, no Babinski sign obtained. Fibrillary twitching is present in the muscles of both legs, in both the wasted and the apparently non-wasted muscles. Sensations are normal to pain, but hypesthesia to touch is found, and some delay is noticed in the temperature sense on each foot, but above the ankles it is normal.

²Review de Méd., 1890.

The upper extremities present some flabbiness in the muscles of the arms, but apparently no atrophy. The muscles of the first interosseous space of each hand are atrophied, also in a less degree the thenar and the hypothenar muscles of each hand. There is also ataxia for fine movements in each hand; and the patient buttons and unbuttons his clothes with difficulty. There is



Photograph of the patient, showing especially the emaciation of the lower limbs.

also some diminution of power in the hands. Sensations of the upper extremities are normal. The sphincters of the bladder and rectum are normal. Sexual weakness has existed for the last two or three years.

The pupils are equal, they react normally to light and in accommodation. No Argyll-Robertson pupil is found. The eye-grounds show an incipient degeneration of both optic nerves. Nystagmus is distinctly present when the eyes are turned to the right, but only slightly when turned to the left. The personal as well

as the family history is negative. He denies venereal diseases and alcohol. He indulged considerably in tea and coffee; the average daily quantity used to be about one hundred ounces a day of tea, and forty ounces of strong coffee.

He complains of pain and numbness in both feet. The electrical examination gave the following result:

The peroneus longus, tibialis anticus, extensor hallucis longus muscles present distinct R. D. under galvanic current, and no response to the faradic current.

The extensor digitorum brevis, and interossei pedis dorsalis muscles show that the CaCC=AnCC on the right side, and CaCC only slightly greater than the AnCC on the left side for the galvanic current, and marked diminution for the faradic electricity. The gastrocnemius on the right side gives very little response to the faradic and galvanic currents, but no R. D.; on the left side slight diminution of electrical contractility. The adductor muscles, the sartorius and quadriceps on either side present diminution of contractility to both currents. The first interosseus space on either side presents very marked diminution of faradic contractility and CaCC is but very slightly greater than the AnCC. The thenar and hypothenar muscles also present slight diminution of electrical contractility.

To sum up, this is a case of progressive muscular atrophy, affecting mostly the peroneal group of muscles. It also shows an involvement of some small muscles of the hands, thus reminding one of Aran-Duchenne's disease; it also reminds one of the pseudo-hypertrophic myopathy by the consistence and appearance of the calf muscles. The muscles which are distinctly wasted show marked diminution of electrical contractility and partial R. D. Total loss of deep reflexes in the lower extremities, fibrillary twitching of the affected muscles, nystagmus and incipient degeneration of the optic nerves, complete the clinical picture of the case. As a coincidence, or perhaps as a cause and effect, I mention the excessive indulgence in tea and coffee.

In comparing this case with those recorded in the literature as primary neurotic atrophy, we find here also an involvement of the upper extremities in addition to that of the peroneal group of muscles. There is also the equino-varus of the feet, the fibrillary twitching, and the complete or partial R. D.

As to the deep reflexes, although in some cases they were found to be exaggerated, in others however they were diminished or lost. My case therefore can be placed alongside of the others with the same symptomatology.

There are, however, a few points which deserve attention. The history gives no trace of a similar disease in any member of the patient's family, while one of the characteristic features of the affection is the existence of the disease in the family. The age at which the disease begins is usually earlier than in my case. Finally, the presence of the nystagmus and changes of the optic nerves are rather unusual.

On the other hand, the equino-varus, the scoliosis, the total loss of deep reflexes, ataxic gait and nystagmus are symptoms characteristic of Friedreich's disease. Finally, the condition of the eye-grounds, the absence of knee-jerks and tendo-Achillis reflex, the paresthetic disturbances in the lower extremities, and the marked impairment of the sexual function, will direct our mind toward tabes.

The case is instructive from the point of view that the line of demarcation between various forms of progressive muscular atrophy cannot be maintained. The latter disease comprises a certain number of types which may differ from each other in the etiology and in the clinical expressions, but they merge into one another and have one underlying basis, namely, a degeneration of the motor neurone, or only of one portion of it, and of the muscular fibers in which this neurone ends. None of the forms possess characteristic features, which would be considered as pathognomonic.

The resemblance of the case to tabes is not unusual, as the type of muscular atrophy under consideration may be accompanied by tabetic symptoms. There are on record cases in which in addition to the degeneration of the anterior roots and anterior horns a degeneration of the posterior roots and columns also was found.

A peculiarity of the case lies also in the fact that tea and coffee were used to a large extent.

In April, 1901, I³ exhibited before this Society a case of tea-intoxication with symptoms of ataxic paraplegia. In discussing the pathogenesis of the cord lesion, I expressed the opinion that the theine, the essential oils of the tea, and the black lead which is used to give a bloom to black tea, are at least the three main ingredients of this beverage that are poisonous to the nervous system. Similar to alcohol, lead and mercury, they are apt to pro-

³Therapeutic Gazette, July, 1901.

duce spinal symptoms or symptoms of diseases of peripheral nerves. If not directly, they may produce a toxic condition, so as to predispose the peripheral or the central nervous system to various diseases.

The disease from which my patient is suffering began with a neuritis, as he experienced severe pain in the toes. The being obliged to stand in a grocery store in cold weather, a work which he had been doing for over twenty years, is not sufficient to cause a degeneration of the peripheral neurones, as examples of exposure without a neuritis are abundant. A special predisposition or vulnerability of the nerve elements is necessary. Is it not likely that the poisons of the tea and coffee which the patient absorbed in large quantities for a number of years, exercised a degenerative effect upon the nervous system and produced the spinal-neurotic atrophy he presents?

The rôle of intoxications (A. Gordon⁴) of any sort upon the nervous system is an established fact, but all the cases reported have reference to mineral poisons, while the effect of the beverages we are absorbing daily as articles of food has not yet been studied in all its phases. The importance of recognizing such a potent element as a causative factor in organic diseases of the nervous system has not yet been adequately realized. The therapist will perhaps be able to draw some practical conclusions from the history of this case.

I wish to express my indebtedness to Dr. John Wanamaker 3d for his assistance in taking the history of the case, and to Dr. Arnold for the photograph.

⁴"The Rôle of Intoxications in Diseases of the Spinal Cord." Phila. Med. Jour., 1901.